Nasal Masses In Children

Abstract
Children presenting with nasal masses in rare. A diagnosis is made easier if the clinician understands their classification and presentational symptoms. The presenting symptoms and signs are discussed together with the principles of management.

Keywords
Nasal Masses, Children.

Introduction
Nasal masses in children are rare but can cause much diagnostic uncertainty. We present a classification of paediatric nasal masses and discuss the principles of assessment and treatment.

Classification
There are a wide range of causes of nasal masses in children. They may be congenital or acquired and can be categorized as either: malignant or benign, midline or lateral, cystic, solid or mixed.

Epidemiology
Congenital nasal masses are rare malformations and occur in one in 20,000-40,000 live births in the USA. There is also a regional variation with fronto-ethmoidal encephaloceles being more common in Southeast Asia, occurring in one in every 5000 births. Therefore due to their rarity, the general Ear Nose and Throat (ENT) surgeon will have little direct experience of these masses.

Presentation
Presentation varies with age and cause. They are rarely detected by pre-natal scanning unless part of a larger mass or where there is extensive pathology e.g. a large intracranial anomaly.

In newborns there may be a visible mass projecting from the nasal cavity. They can present as an emergency with respiratory distress due to the fact that newborns tend to be obligate nose breathers. They may become cyanotic on feeding or on crying -cyclical cyanosis. There may be difficulty in passing a suction catheter or nasogastric tubing. An external deformity e.g. splaying of the bones of the mid-face or an obvious bone or soft tissue swelling can also be seen.

In older children the symptoms may be more varied. Epistaxis, nasal obstruction, rhinosinusitis and deafness due to otitis media with effusion (OME) have all been described. However the most common presenting complaint of a nasal mass to the ENT department is with nasal obstruction.

Management
Neonates may present with respiratory distress due to complete nasal obstruction. The management starts with securing a safe airway. Following this, a full history and examination should be taken. Prior to any biopsy or aspirate it is imperative to perform imaging. This is not just to show any possible intracranial connection but to demonstrate the nature and extent of the mass.

Assessment
History
A full paediatric ENT history will need to be taken from the parents and carers and also from the patient if possible. However in certain cases, the airway may need to be secured prior to this. The history should start with the birth history including prematurity, mode of delivery and the APGAR scores.

Clinical Examination
A thorough examination of the upper respiratory system and head neck region should be carried out. General inspection may reveal the patient to only mouth breath or show evidence of increased work of breathing. This
may include tracheal tug or substernal recession. A skin crease in the supra-tip region may indicate frequent nose rubbing. Anterior rhinoscopy may reveal any nasal polyps, lesions, nasal discharge, rhinitis or evidence of bleeding. Assessment of a patent nasal airway can be performed using a metal tongue depressor or a laryngeal mirror. The amount of misting may help identify any obvious nasal obstruction and whether it is unilateral or bilateral. Passing a nasogastric tube is often used on the paediatric wards to assess for a patent nasal airway in newborns and help exclude choanal atresia. Either a flexible or rigid nasoendoscope is normally well tolerated in older children if local anaesthesia is used. The endoscope is a useful tool in assessing the nasal passage and post nasal space. Along with identifying any pathology, it can provide reassurance to the patient and family especially if the camera and video screen are used.

Adenoids can cause diagnostic confusion and may be well seen at endoscopy. Adenoid size can be graded and starts with grade I which is the adenoid filling one-third of the vertical portion of the choanae to grade IV which is complete choanal obstruction.2

Oro-pharyngeal examination may reveal a post nasal drip, bulging of the soft palate or even a post nasal mass descending into the oropharynx.

Neck examination should be performed for any neck masses looking in particular at the posterior triangle which is the lymphatic drainage of the post nasal space. There is no intra-cranial connection. An MRI can differentiate between masses containing glial cells, fibrous and vascular tissue. They are similar to encephaloceles but have become separated from the intracranial structures, although some may remain attached with a fibrous stalk.

Imaging
It is mandatory to perform imaging prior to a biopsy or excision of any nasal mass in children due to the possibility of an intracranial connection. This could cause a potential cerebrospinal fluid (CSF) leak or infection such as meningitis or encephalitis. There are also very vascular lesions such as nasopharyngeal angiofibromas which bleed profusely on biopsy and that can be diagnosed on imaging. Ideally the imaging modality should be discussed with a paediatric radiologist so the optimum method can be chosen depending on the clinical need.

Magnetic resonance imaging (MRI) provides detailed imaging of the nose and post nasal space and has the advantage of soft tissue imaging and information on any intracranial connection (Figure 1). A Computed Tomography (CT) scan is sometimes a complementary tool along with ultrasound scanning to assess any associated neck masses. CT scanning is better at assessing any bony defects in the skull base.

Treatment
This very much depends on the suspected diagnosis. A Multi-Disciplinary Team (MDT) meeting will enable the scans to be discussed in detail and plan out any procedure. A multidisciplinary approach may be required for any intracranial connection and a Paediatric Oncologist is urgently needed for the malignant masses.

Congenital Nasal Masses
Congenital midline nasal masses are rare and can be composed of tissue that has herniated from the cranium or may be made of tissue native to the nasal cavity. Encephaloceles, Meningocele and Gliomas can consist of ectopic intracranial tissue and may have an intracranial connection.

Dermoids in contrast are not composed of intracranial tissue and are thought to be due to the failure of the normal separation of the different germ cell layers.

Encephalocele, Meningocele and Glioma
Encephalocele is a herniation of meninges with or without brain tissue through a defect in the calvarium. An encephalocele will retain an intracranial connection but this may have atrophied at the time of diagnosis. A meningocele consists of either only meninges or of meninges with CSF but does not contain nervous tissue. Gliomas are benign midline masses containing glial cells, fibrous and vascular tissue. They are similar to encephaloceles but have become separated from the intracranial structures, although some may remain attached with a fibrous stalk.

Clinical Features
These nasal masses generally present at birth with breathing problems. Nasal encephaloceles are generally found in the root of the nose or inferior to the nasal bones. They are compressible and transilluminate and thus can be confused with nasal polyps.

Nasal gliomas are usually firm and noncompressible masses with a negative Furstenberg’s test.3 Furstenberg’s test (compression of the Internal Jugular Vein) would cause an encephalocele to enlarge but not a glioma. Gliomas can be confused with nasal haemangiomas because they can be covered with telangiectasia and may be purple in colour.

Imaging
Imaging is essential. A CT image is preferred for assessing the bony defect in the skull base while MRI is able to assess for an intracranial connection. An MRI can differentiate between gliomas and encephalocele by demonstrating an associated
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CSF space. Nasal gliomas may contain areas of calcification or cystic changes on CT scan and tend to be isodense on CT. On MRI, gliomas are usually hyperdense on T2-weighted images.

Treatment
Nasal encephaloceles if left untreated can cause CSF leaks, meningitis and intracranial abscess. This is still a risk with gliomas but to a much lesser extent.

After appropriate imaging the management can be planned. Intranasal masses with no intracranial communication can be removed by a transnasal approach, often endoscopically using a microdebrider. External lesions may be excised with an overlying skin incision, an external rhinoplasty approach or lateral rhinotomy for extensive lesions.

In those with intracranial connections or suspected intracranial connections a combined approach is required with the paediatric neurosurgeons.

Nasal Haemangioma
These are solid endothelial tumours which undergo a period of rapid proliferation between the ages of approximately 6 weeks and 12 months followed by involution. During the rapid growth phase propranolol +/- steroids is the first line treatment to control growth. Anti-proliferative drugs may be indicated if the haemangioma is enlarging and causing damage to surrounding structures, particularly the orbit as they can compromise vision. Larger or external lesions may require surgery.

Congenital Teratomas
Teratomas are rare tumours containing components of ectoderm, mesoderm and endoderm. It can be diagnosed on prenatal screening if an elevated α-fetoprotein is picked up but the most common presenting symptom in neonates is respiratory distress. They are more commonly seen in females and generally arise from the midline or lateral nasopharyngeal wall (Figure 2).

Hamartomas
Hamartomas are non-neoplastic malformations. They contain abnormal mixture of tissues indigenous to that area of the body. They are extremely rare but can cause nasal obstruction in newborns.

Hairy Polyps
There is some confusion over the classification of these lesions. They have been previously described as teratomas, hamartomas, dermoids or choristomas. These lesions are most commonly known as teratomas. However by definition, teratomas consist of all three germinal layers and hairy polyps consist of a mesodermal core surrounded by ectoderm.

Hairy polyps in the nasopharynx also present with airway obstruction or feeding problems in children. The treatment is surgical excision and they can be removed transorally and/or endoscopically.

Cystic Nasal Masses

Dermoid Cyst
Dermoid cysts are the commonest midline nasal mass. They derive from ectoderm and mesoderm and can contain all the structures of normal skin (including hair follicles, sweat glands and sebaceous glands).

Clinical Features
A nasal dermoid will usually present as a slow growing cystic midline mass over the dorsum of the nose. An associated pit can often be seen in any position from the nasal tip to the glabella. The cyst can become infected and form an abscess. They are usually diagnosed within the first three years of life. They can cause broadening of the nasal dorsum and deformation of the nasal bones or cartilage.

They are usually firm and do not transilluminate or compress. They demonstrate a negative Furstenberg test (no expansion with increased venous pressure).

Imaging is necessary to assess for any intracranial extension as nasal dermoid cysts have an intracranial connection in about 20% of cases. There are often associated congenital anomalies but no known related syndrome.

Imaging
MRI is both sensitive and specific and is better at visualizing the soft tissues and diagnosing intracranial extension. Nasal dermoids appear hyperdense on T1-weighted MRI.

CT scanning is useful for visualising bony defects of the skull base. A bifid crista galli and widened foramen cecum suggest intracranial involvement of the dermoid. However 14% of children under the age of 1 year will have incomplete ossification of these areas.

Management
In general dermoids cysts should be excised before they cause complications. Complications can include local infection, inflammation or abscess formation. An intracranial connection may result in CSF leakage, meningitis, cavernous sinus thrombosis or periorbital cellulitis.

The surgical approach depends on the size and position of the cyst and whether there is any intracranial connection. This needs to be decided following the necessary imaging. The nasal portion of the dermoid can be removed using a midline vertical, transverse, lateral rhinotomy or midbrow

Figure 2: With the soft palate retracted a nasal teratoma can be seen filling the post nasal space.
incision with acceptable scarring and good cosmetic results. An open rhinoplasty approach may provide enough access in a cyst in the lower half of the nose and closed rhinoplasty has also been used. A lesion higher up in the nose would require direct excision and any intracranial extension would necessitate a combined approach. Dorsal reconstruction may be required with a cartilaginous graft. More recently endoscopic approaches have been used.

**Nasolacrimal Duct Cysts**

If the lumen of the nasolacrimal duct is blocked then fluid can build up forming a cyst. It can cause epiphoria and nasal obstruction. The majority resolve spontaneously by nine months of age.

**Nasalveolar Cysts**

These arise from the incisive canal during the development of the maxilla. They present lateral to the midline at the alar base.

**Thornwaldt’s Cyst**

They are the result of the primitive notochord remaining connected to the nasopharynx with the development of a bursa or pouch. The cysts are generally smooth, submucosal masses and located superior to the adenoidal pad.

**Clinical Features**

The majority of Thornwaldt cysts present in the second or third decade as a midline nasopharyngeal mass.

**Imaging**

An MRI scan will show any adhesion to the cervical vertebrae. They have high signal intensity on T1 and T2 weighted images.

**Management**

The treatment is generally with primary surgical resection.

**Dentigerous Cysts**

These are cysts with a dental origin (odontogenic). A well demarcated radiolucent lesion is seen on x-ray adjacent to an unerupted tooth. The common sites for dentigerous cysts are the mandibular 3rd molars and the maxillary canines.

**Mucous Cysts**

These are mainly extranasal masses. These may rarely occur anywhere within the nose but more commonly along the floor of the nose.

**Nasal Tumours**

Nasal tumours in children are rare. They may be benign or malignant and can be derived from epithelial, lymphatic or connective tissue.

**Juvenile Nasopharyngeal Angiofibroma**

Nasopharyngeal angiofibromas is a benign highly vascular tumour. It is the commonest type of benign nasopharyngeal tumour and accounts for about 0.5% of all head and neck tumours. The highest reported occurrence is in India and Egypt and they most commonly occur in prepubescent boys.

**Clinical Features**

The most common presenting symptoms are nasal obstruction and epistaxis. Although they originate in the posterior nasal cavity by the time of presentation they have grown to fill the nasopharynx. They are typically seen in boys and the mean age of presentation is 14 years old. If untreated, they can cause upper airway obstruction and progressive intracranial involvement.

**Imaging (Figure 3)**

On imaging these tumours arise on the posterolateral wall of the nasal cavity at the level of the sphenopalatine foramen. They tend to extend into the pterygopalatine foramen. On CT the tumour is multilobulated and enhances with contrast.

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**Figure 3:** Contrast enhanced T1W scans in axial and coronal planes demonstrating a nasopharyngeal angiofibroma.
It can show bone invasion but classically causes bowing of the posterior wall of the maxillary sinus due to the size of the tumour. On MRI the lesion is hypervascular but the lesion may also be heterogeneous due to cyst formation and areas of haemorrhage. On T1-weighted images they appear hypodense and moderately hyperdense on T2-weighted images.

Treatment
The tumour growth rate is slow and due to the fact they are not seen in adults it is thought they regress spontaneously. The primary treatment is surgical resection. This can be performed endoscopically for small lesions and the majority can be successfully managed with less invasive techniques.16 Other surgical approaches for resection include a lateral rhinotomy and medial maxillectomy approach. In large lesions a combined approach may be required. If complete excision is not possible then radiotherapy has been shown to be effective at controlling the symptoms.17 To excise these vascular tumours pre-operative embolization and hypotensive anaesthesia are often used. This decreases the amount of blood loss and enables large tumours to be excised. There are risks with embolization of causing accidental embolization to cerebral and ophthalmic arteries.

Rhabdomyosarcoma
Rhabdomyosarcoma account for 60% of all sarcomas in the paediatric population and 40% occur in the head and neck.18 Nearly half of these occur in children under 5 years old. The common sites include the orbit, nasopharynx, sinus and middle ear.

There are different histological subtypes which are important in the treatment and prognosis. The most common subtype is embryonal. Alveolar and undifferentiated sarcomas are associated with a worse prognosis.

Signs and Symptoms
It may present with nasal obstruction, a firm posterior triangle node or unilateral eustachian tube dysfunction. Horner’s syndrome and cranial nerve palsy are late signs when the tumour has become more advanced.

Management
Following detailed imaging and biopsy treatment can be planned. There are guidelines for treatment according to the Intergroup Rhabdomyosarcoma Studies.19 Primary surgical resection may be possible in localised disease but often these tumours present late. Therefore chemo-radiotherapy with or without surgery is the main treatment option.

Lymphoma
In Asian populations sinonasal lymphomas are second only to gastrointestinal lymphomas as the commonest type of extranodal lymphomas. They can be separated into B-cell, T-cell and natural killer cell phenotypes. Lymphoepitheliomas and lymphosarcomas also occur but are rare.

Nasopharyngeal Carcinoma
Nasopharyngeal carcinoma is an uncommon tumour accounting for only 1-2% of paediatric malignancies in the
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UK and US. However in other geographical locations such as parts of Africa, 10-20% of childhood malignancies are due to nasopharyngeal carcinoma. There is a bimodal bidistribution of this disease with an early peak of 10-20 years and a second peak between 40-60 years. There is an association with Epstein-Barr virus. Males are more commonly affected.

The majority of patients with nasopharyngeal carcinoma will present with metastatic disease in the neck. Other common presenting symptoms include otitis media, rhinorrhea and nasal obstruction. Cranial nerve palsy or facial pain would suggest skull base invasion. Surgical resection may not be possible with advanced disease and chemo-radiotherapy would be the main treatment.

Olfactory Esthesioneuroblastoma

These rare tumours arise from the olfactory epithelium. They are initially unilateral but can grow into the adjacent sinuses and the contralateral nasal cavity. They can also invade into the orbit and brain. Management is with combined modality treatment. The five-year disease free and survival is 68%. The best survival rates are with combined surgery and radiation.

Fibro-osseous Disease

Fibro-osseous lesions vary in there clinical behaviour and radiological features. Aggressive lesions require a radical surgical approach to ensure complete excision. Incomplete excision of aggressive lesions may result in disease recurrence with severe morbidity or mortality. In contrast a slowly progressive lesion often does not warrant extensive surgical excision.

Foreign bodies and rhinoliths

Nasal foreign bodies can present as a nasal mass. A long standing foreign body is thought to cause a Rhinolith. The foreign body becomes encrusted by mineral deposits. As they become larger they cause nasal obstruction and unilateral rhinorrhea. CT scanning can reveal bony erosion of the floor of the nose. In some cases the rhinoliths have been so large they require a lateral rhinotomy to excise them.

Nasal Polyps

Nasal polyps are not common in children but can occur in severe allergic rhinitis. Children with cystic fibrosis are also at greater risk and an unexplained nasal polyp in a child should prompt a screening test for cystic fibrosis.

References


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